



Editorial

The Current Strategy in Managing Congenital and Structural Heart Disease: Find and Treat Them Appropriately

Radityo Prakoso^{1*}

¹Division of Pediatric Cardiology and Congenital Heart Disease, Department of Cardiology and Vascular Medicine, National Cardiovascular Centre Harapan Kita, Universitas Indonesia, Jakarta, Indonesia

ARTICLE INFO

Keyword :
Fetal Intervention;
Fontan;
Non-Fluoroscopy;
Percutaneous;
Reverse Potts;
Rheumatic Heart Disease;
RVOT Stenting.

ABSTRACT

Significant progress has been made in diagnosing and treating congenital heart defects (CHDs) over the past five decades. Notable advancements include pulse oximetry screening for critical CHDs in newborns and percutaneous, catheter-based procedures, improving outcomes even in developing nations. Recent studies demonstrate favorable outcomes with device closure of acyanotic CHDs, with non-fluoroscopy techniques adopted to minimize radiation risks. Positive results are observed with right ventricular outflow tract (RVOT) stent palliation for late-presenting cyanotic congenital heart diseases such as tetralogy of Fallot (TOF). Novel interventions like reverse Potts shunt anastomosis show promise in managing pediatric pulmonary arterial hypertension (PAH), while prophylactic treatments effectively reduce postoperative arrhythmias. Fontan procedure significantly enhances survival rates for individuals with single functional ventricles, although challenges arise with aging populations. In developed countries, fetal cardiac interventions utilizing advanced imaging techniques allow for early detection and preventive interventions, reducing secondary complications.

Over the past five decades, significant progress has been made in diagnosing and treating congenital heart defects (CHDs). Notable advancements include the introduction of pulse oximetry screening for critical CHDs in newborns prior to hospital discharge, alongside traditional diagnostic methods such as history-taking, physical examination, chest X-rays, and electrocardiograms. The utilization of percutaneous, catheter-based procedures and advanced surgical techniques has greatly improved outcomes, even in developing nations.¹⁻³

Recent studies have demonstrated favorable outcomes with device closure of acyanotic CHDs, including atrial septal defect (ASD), ventricular septal defect (VSD), and patent ductus arteriosus (PDA).^{4,7} Concerns over radiation exposure, especially in children and pregnant women, have led to the adoption of non-fluoroscopy techniques for defect closure, with comparable efficacy and safety to traditional methods. In our effort to mitigate radiation exposure during transcatheter closure procedures for congenital heart defects, we have implemented a policy promoting non-fluoroscopy techniques, even in challenging cases such as multiple ventricular septal defects (VSDs) and in pregnant women with atrial septal defect (ASD) with pulmonary hypertension.^{4,5} This strategic shift aims to minimize the potential risks associated with radiation exposure, particularly for vulnerable populations. In one study demonstrated by Siagian et al.,⁶ echocardiography-guided closure of patent ductus arteriosus (PDA) is a viable alternative to fluoroscopy-guided procedures. Their study, involving 30 patients who underwent non-fluoroscopy closure, revealed that echocardiography-guided closure is comparable in efficacy to fluoroscopy-guided closure, despite potential limitations in visualization. Importantly, the procedural time for echocardiography-guided PDA closure did not differ significantly from fluoroscopy-guided closure, indicating that reliance solely on echocardiography did not compromise procedural efficiency.

Another problem in developing country is rheumatic heart disease (RHD), often leading to mitral stenosis (MS). Percutaneous balloon mitral valvotomy (PBMV) has emerged as a preferred treatment over surgical options due to its superior outcomes and shorter hospital stays. However, challenges persist, particularly in special populations such as pregnant women and those with arrhythmias like atrial fibrillation.⁸ Late presentations cases in cyanotic congenital heart disease such as tetralogy of Fallot (TOF) are also common in developing countries, posing complex challenges. Recent studies suggest positive outcomes with right ventricular outflow tract (RVOT) stent palliation, leading to improvements in oxygen saturation and ventricular function. Despite advancements in diagnostic technologies and a deeper comprehension of the ailment compared to previous centuries, instances of delayed presentations persist. Late presentations are particularly prevalent in developing countries, exacerbating the complexity of the issues. Traditionally, patients with uncorrected Fallot physiology who present late were solely managed conservatively with medication, especially when significant desaturation and diminished ventricular function were present. We demonstrated that there was a significant improvement in patients' saturation, rising from an initial mean of $58.56\% \pm 19.03\%$ to $91.03\% \pm 8.98\%$ ($p < 0.001$), along with an increase in their left ventricular ejection fraction (LVEF) from $64.00\% \pm$

* Corresponding author at: Department of Cardiology and Vascular Medicine, National Cardiovascular Centre Harapan Kita, Universitas Indonesia, Jakarta, Indonesia

E-mail address: karajanh70@gmail.com (R. Prakoso).

<https://doi.org/10.21776/ub/hsj.2024.005.02.2>

Received 10 March 2024; Received in revised form 13 March 2024; Accepted 15 March 2024.

Available online 28 April 2024

18.21% to 75.09% \pm 12.98% ($p = 0.001$). Notably, three patients experienced substantial enhancements in their LVEF, with values rising from 31% to 55%, 31% to 67%, and 26% to 50%. The median duration of hospital stay was 8 days (range: 2-35 days), with a median intensive care unit (ICU) stay of 2 days (range: 0-30 days). The median interval from right ventricular outflow tract (RVOT) stent palliation to total repair was 3 months (range: 1-12 months).^{3,9}

Pediatric pulmonary arterial hypertension (PAH) remains a progressive disease with limited treatment options. Novel interventions, such as the reverse Potts shunt anastomosis, show promise in converting severe PAH to a more manageable condition.¹⁰ Junctional Ectopic Tachycardia (JET) is a common postoperative arrhythmia, particularly following congenital heart surgery. Prophylactic treatments, including dexmedetomidine, amiodarone, and magnesium sulfate, have shown efficacy in reducing JET incidence, with dexmedetomidine potentially offering additional benefits.¹¹ Fontan procedure remains a cornerstone treatment for individuals with single functional ventricles, boasting enhanced survival rates over the years. Following the inception of the Fontan technique, the 10-year survival rate post-surgery has surged from a mere 50% to its present 90%. This procedure has significantly elevated survival prospects for individuals born with single ventricles. Nonetheless, as this population ages, the physiological challenges become more apparent. Bosentan has exhibited promise in enhancing exercise tolerance and overall quality of life among Fontan patients, although additional research is warranted to refine treatment approaches.¹²

In developed countries, fetal cardiac intervention and surgery have become increasingly common. With advanced fetal imaging techniques like echocardiography and magnetic resonance imaging, CHDs can often be detected as early as the 12th week of gestation. Early intervention during fetal development can prevent secondary cardiac complications, such as ventricular hypoplasia. Fetal interventions, particularly for conditions like fetal aortic stenosis and pulmonary atresia with an intact ventricular septum, have shown promise.^{13,14}

Conflict of Interest

There is no conflict of interest.

References

- Baumgartner H, Backer JD, Babu-Narayan SV, et al. 2020 ESC guidelines for the management of adult congenital heart disease. *Eur Heart J*. 2021; 42: 563-645. doi: 10.1093/eurheartj/ehaa554
- Stout KK, Daniels CJ, Aboulhosn JA, et al. 2018 AHA/ACC guideline for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association task force on clinical practice guidelines. *Circulation*. 2019;139:e698–e800. doi: 10.1161/CIR.0000000000000603
- Prakoso R, Kurniawati Y, Siagian SN, Sembiring AA, Sakti DDA, Mendel B, Pratiwi I, Lelya O, Lilyasari O. Right ventricular outflow tract stenting for late presenter unrepaired Fallot physiology: a single-center experience. *Front. Cardiovasc. Med.* 2024;11:1340570. doi: 10.3389/fcvm.2024.1340570
- Prakoso R, Ariani R, Mendel B, Lilyasari O. Transoesophageal echocardiography-guided balloon-assisted percutaneous closure of a large secundum atrial septal defect in a pregnant woman: a case report. *Eur Heart J Case Rep.* 2024 Jan 6;8(1):ytae014. doi: 10.1093/ehjcr/ytae014. PMID: 38274706; PMCID: PMC10810588.
- Siagian SN, Prakoso R, Mendel B, Hazami Z, Putri VYS, Zulfahmi, Sakti DDA, Kuncoro AS. Transesophageal echocardiography-guided percutaneous closure of multiple muscular ventricular septal defects with pulmonary hypertension using single device: A case report. *Front Cardiovasc Med.* 2023 Mar 23;10:1093563. doi: 10.3389/fcvm.2023.1093563. PMID: 37034333; PMCID: PMC10076841.
- Siagian SN, Prakoso R, Putra BE, Kurniawati Y, Lelya O, Sembiring AA, Atmosudigdo IS, Roebiono PS, Rahajoe AU, Harimurti GM, Mendel B, Christianto C, Setiawan M, Lilyasari O. Echocardiography-Guided Percutaneous Patent Ductus Arteriosus Closure: 1-Year Single Center Experience in Indonesia. *Front Cardiovasc Med.* 2022 May 23;9:885140. doi: 10.3389/fcvm.2022.885140. PMID: 35677684; PMCID: PMC9167953.
- Mendel B, Amin BF, Prakoso R. Potency of non-fluoroscopy guided patent ductus arteriosus closure: a case report. *SN Compr Clin Med* 2021; 3: 2665-72
- Habib F, Mendel B, Fauzan R, Nasution AN. Revisiting percutaneous balloon mitral valvotomy technique and safety in various population: an evidence-based case report and literature review. *Front Cardiovasc Med.* 2024 Jan 26;11:1334444. doi: 10.3389/fcvm.2024.1334444. PMID: 38343874; PMCID: PMC10853446.
- Prakoso R, Agita Sembiring A, Dwisepto Aulia Sakti D, Mendel B, Lilyasari O. Double stenting in 19-year-old patient with tetralogy of Fallot with prior fractured stent. *J Am Coll Cardiol Case Rep.* 2022;4(20):1375–8. doi:10.1016/j.jaccas.2022.07.043
- Mendel B, Christianto C, Angellia P, Holiyono I, Prakoso R, Siagian SN. Reversed Potts Shunt Outcome in Suprasystemic Pulmonary Arterial Hypertension: A Systematic Review and Meta-Analysis. *Curr Cardiol Rev.* 2022;18(6):e090522204486. doi: 10.2174/1573403X18666220509203335. PMID: 35538823; PMCID: PMC9893152.
- Mendel B, Christianto C, Setiawan M, Prakoso R, Siagian SN. A Comparative Effectiveness Systematic Review and Meta-analysis of Drugs for the Prophylaxis of Junctional Ectopic Tachycardia. *Curr Cardiol Rev.* 2022;18(1):e030621193817. doi: 10.2174/1573403X17666210603113430. PMID: 34082685; PMCID: PMC9241111.
- Mendel B, Christianto, Setiawan M, Siagian SN, Prakoso R. Pharmacology Management in Improving Exercise Capacity of Patients with Fontan Circulation: A Systematic Review and Meta-analysis. *Curr Cardiol Rev.* 2022;18(5):34-49. doi: 10.2174/1573403X18666220404101610. PMID: 35379155; PMCID: PMC9896416.
- Mendel B, Kohar K, Amirah S, Vidya AP, Utama KE, Prakoso R, Siagian SN. The outcomes of fetal aortic valvuloplasty in critical aortic stenosis: A systematic review and meta-analysis. *Int J Cardiol.* 2023 Jul 1;382:106-111. doi: 10.1016/j.ijcard.2023.03.050. Epub 2023 Mar 29. PMID: 36996909.
- Mendel B, Kohar K, Yumnanisha DA, Djiu RJ, Winarta J, Prakoso R, et al. Impact of fetal pulmonary valvuloplasty in in-utero critical pulmonary stenosis: A systematic review and meta-analysis. *Int J Cardiol CHD.* 2024; 15(2024): 100485